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CASE REPORT

Cor Triatriatum in an Adult with Late Presentation of Symptoms

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KEY WORDS
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Cor triatriatum is a rare congenital heart disease characterized by the presence of a membrane that divides the left atrium into posterosuperior and anteroinferior chambers. Classically, patients are diagnosed in childhood. We report an adult case of cor triatriatum presenting with dyspnea, and note that the strengthened use of transthoracic echocardiography study helps the differential diagnosis of patients complaining of dyspnea and orthopnea in the emergency department.

Introduction

Cor triatriatum is a rare congenital heart disease characterized by the presence of a membrane which divides the left atrium into two chambers. The clinical features mimic those of mitral stenosis. The use of transthoracic echocardiography study in the emergency department helps the differential diagnosis of the patients complaining of dyspnea and heart failure symptoms promptly and provides clues for further management and disposition correctly.

Case report

A 53-year-old man presented to the emergency department due to orthopnea and progressive dyspnea over the past week. He was a builder with good exercise tolerance. He had a blood pressure of 141/76 mmHg and an irregular rapid pulse rate of 156 beats/minute. He had a respiratory rate of 20 breaths/minute and an oxygen saturation of 96%. Physical examinations revealed engorged jugular vein and bilateral rales over bilateral basal lung area without audible cardiac murmur. The results of the remainder of his physical examination were not remarkable. Electrocardiogram showed atrial fibrillation with rapid ventricular response (Fig. 1). Chest roentgenogram revealed cardiomegaly with tortuous aorta. No active pulmonary lesion was found (Fig. 2). Laboratory studies were all within normal limits.

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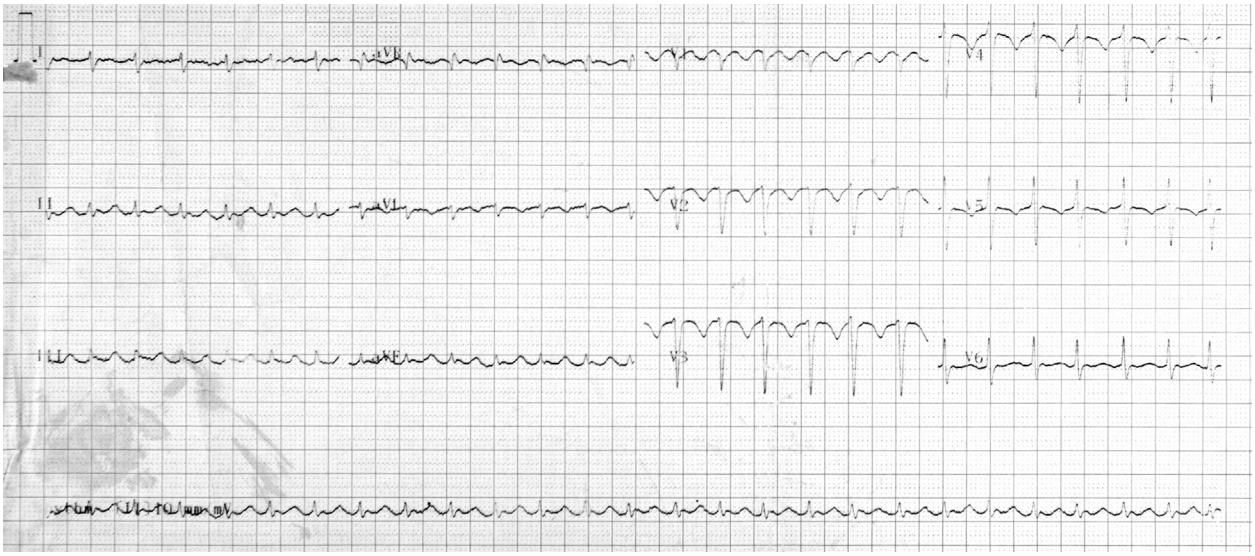

Fig. 1 Electrocardiogram shows atrial fibrillation with rapid ventricular response.

Transthoracic echocardiography in the emergency department showed the presence of a membrane that divides the left atrium into posterosuperior and anteroinferior chambers membrane from the apical four chamber view (Fig. 3). A dilated left ventricular chamber and an impaired left ventricular contractility were also found. Magnetic resonance imaging provided visualization of the anatomical correlation of these two left atrial chambers and pulmonary veins (Fig. 4). This patient received surgical excision of the left atrial membrane and was discharged on the 10th day after the operation with good recovery. Follow-up transthoracic echocardiography of this patient showed improved left ventricular contractility and decreased mitral regurgitation.

Discussion

Cor triatriatum is a rare congenital heart disease first described by Church in 1868 [1]. Cor triatriatum was reported to have an incidence of 0.4% at autopsy of patients with congenital cardiac disease and 0.2% among patients

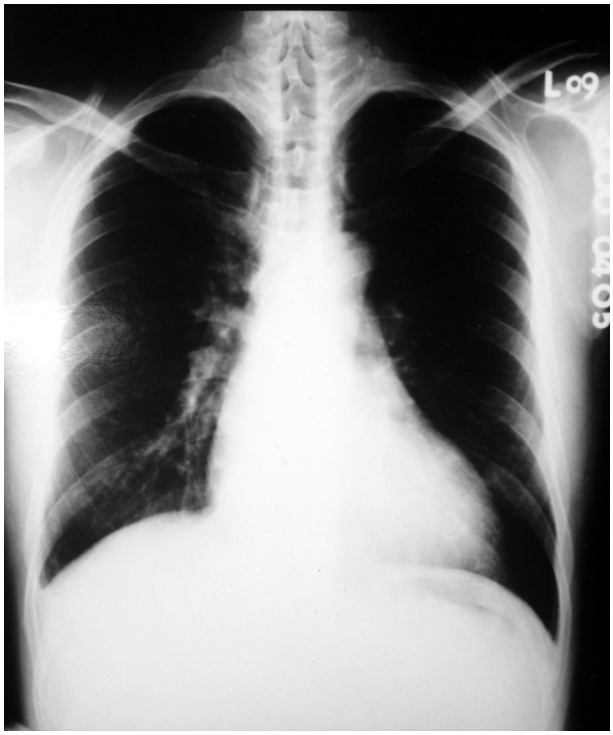

Fig. 2 Chest roentgenogram reveals cardiomegaly with tortuous aorta. No active pulmonary lesion is found.

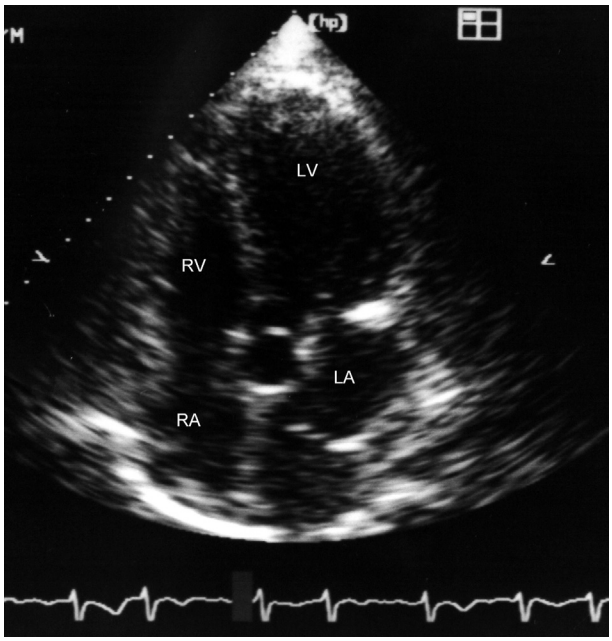

Fig. 3 Transthoracic two-dimensional echocardiography shows the presence of a membrane that divides the left atrium into posterosuperior and anteroinferior chambers from the apical four chamber view.

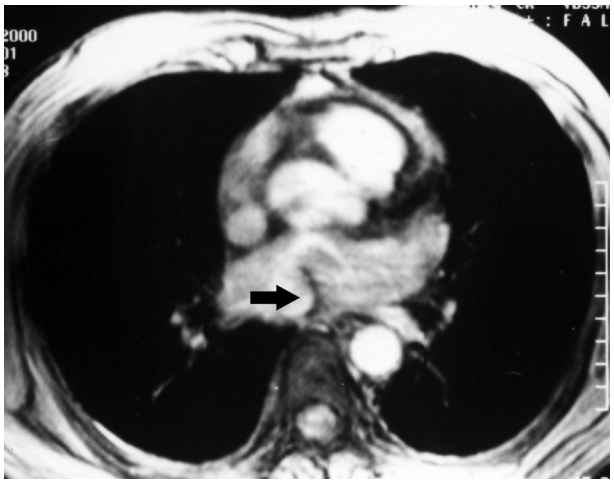

Fig. 4 Magnetic resonance imaging reveals the long-axis view of the intra-atria fibrous membrane (arrow) in the left atrium.

undergoing transesophageal echocardiography. The incidence of cor triatriatum is less than 1 in 10,000 in high-volume echocardiographic laboratories [2]. Cor triatriatum is characterized by the presence of a membrane that divides the left atrium into two chambers: a posterosuperior chamber receiving the pulmonary veins, and an anteroinferior chamber communicating with the mitral orifice [3]. The membrane within the left atrium results from a failure of reabsorption of the common pulmonary vein during development and then hinders the pulmonary venous flow [4]. The clinical features mimic those of mitral stenosis [5], and the degree of obstruction depends on the diameter of the orifice between the two portions of the left atrium. This disease is usually diagnosed in childhood, although sometimes it is also found in adults [4]. Cor triatriatum in adults has been reported in association with atrial septal defect and dilated coronary sinus due to persistent left superior vena cava, and bicuspid aortic valve [6,7]. The development of atrial fibrillation or mitral regurgitation may be responsible for these late clinical presentations [5]. This patient remained undiagnosed until the meridian of life, and the concurrent atrial fibrillation disclosed in the emergency department may result in his presentation of heart failure.

Transthoracic echocardiography is an optimal initial examination in the diagnosis and assessment of the disease. Because of its easy utility and ready availability, transthoracic echocardiography has become an important modality for emergency physicians to evaluate patients presenting with dyspnea, chest pain, and cardiac arrest. It allows clinical physicians to differentiate cor triatriatum from mitral stenosis, pulmonary vein stenosis, and acute coronary syndrome. It also reveals the degree of the obstruction that exists. Furthermore, echocardiography is useful in the differential diagnosis of other associated congenital heart diseases [6,7]. The transthoracic echocardiography of this patient showed a typical membrane dividing the left atrium into two chambers with moderate mitral regurgitation. Magnetic resonance imaging provides visualization of the anatomical correlation and is one of the gold standards in the assessment of congenital heart diseases [8]. In this patient, it provided visualization of the anatomical correlation of these two left atrial chambers and pulmonary veins.

Medical care in symptomatic patients includes hemodynamics stabilization, management of fluid overload and pulmonary edema, control of ventricular response and anticoagulation in patients with atrial fibrillation, and anticoagulant prophylaxis in patients with right heart failure. In symptomatic patients, surgical excision of the left atrial membrane usually leads to good recovery [9,10]. The common approach is complete resection of the membrane and closure of the atrial septum. Correction of associated congenital defects also needs to be performed. In this patient, corrected mitral regurgitation and decreased ventricular response of atrial fibrillation after surgical intervention may result in the improvement of left ventricular contractility.

In summary, the use of transthoracic echocardiographic study in the emergency department facilitates the prompt differential diagnosis of patients complaining of dyspnea and heart failure symptoms and thus correctly provides clues for further management and disposition of the patient.

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